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Rare cause of haemoptysis: bronchopulmonary sequestration

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SUMMARY

Bronchopulmonary sequestration is a rare congenital pulmonary abnormality of the lower airways, which includes an abnormal and non-functioning lung tissue not communicating with the tracheobronchial tree and having aberrant blood supply from systemic circulation with variable venous drainage. The incidence of sequestration is around 0.15%–6.4% of all congenital lung malformations. Common presenting features are cough and expectoration. Misdiagnosed cases may present with recurrent infections and haemoptysis. CT of the chest with contrast is the imaging modality of choice. This is a case report of a 32-year-old woman who presented with cough and haemoptysis. CT of the chest showed a multiloculated mass-like lesion in the left lower lobe with a feeding artery from coeliac plexus and venous drainage via the normal left pulmonary vein. Based on CT chest findings, diagnosis of intralobar pulmonary sequestration was made. The patient was reviewed by cardiothoracic surgeons and underwent surgical resection of the sequestered lung. Common presenting features are cough and expectoration. Misdiagnosed cases may present with recurrent infections and haemoptysis. CT of the chest with contrast is the imaging modality of choice.

BACKGROUND

Bronchopulmonary sequestration is divided into intralobar and extralobar types.¹ The most common type is intralobar sequestration, and it is commonly located in the lower lobes of the normal lung and does not have its pleural covering. Intralobar subtype comprises 75% of the pulmonary sequestration, and the incidence is similar in men and women.^{2,3}

On the contrary, extralobar sequestration is situated outside the normal lung with its pleural covering. It constitutes about 25% of bronchopulmonary sequestrations,³ is generally associated with other congenital anomalies and is often diagnosed at an early age.⁴

This is a case report of intralobar pulmonary sequestration in a 32-year-old woman.

CASE PRESENTATION

A 32-year-old woman was admitted with a 1-week history of moderate haemoptysis associated with vague chest pain and chest tightness. There were no associated symptoms, including fever, weight loss, loss of appetite and dyspnoea.

She underwent thyroidectomy in the past, but she never had a chest X-ray before this admission.

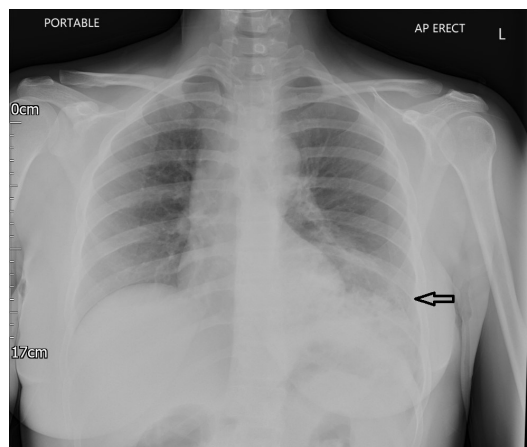


Figure 1 Chest X-ray on admission.

On examination, air entry was noticed to be decreased at the left base and oxygen saturation was 97% on room air. Initial blood results showed marginally raised white cell count with neutrophilia and mildly raised C reactive protein.

Chest X-ray on admission revealed heterogeneous opacity on the left lower zone (figure 1). Ultrasound of the abdomen and pelvis was normal. Based on the above information, an initial diagnosis of community-acquired pneumonia was made and she was started on empirical antibiotics as per local trust protocol. However, she continued to have intermittent mild haemoptysis.

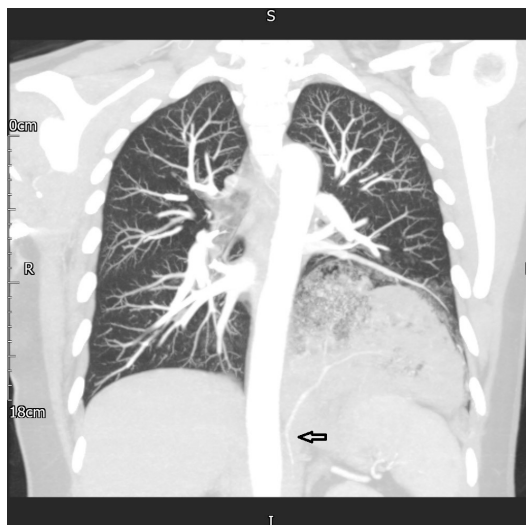


Figure 2 CT of the chest with contrast (lung window) demonstrating the feeding vessel into the sequestered lung.



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Figure 3 CT of the chest with contrast (mediastinal window) demonstrating the feeding vessel from coeliac plexus into the sequestered lung.

Given the suboptimal response to the treatment and drop in haemoglobin from 129 g/L to 103 g/L, CT of the chest with contrast was done, which showed a multiloculated cystic solid mass with low-density lesions and a feeding vessel from the coeliac artery, suggestive of intralobar bronchopulmonary sequestration (figures 2–6). Ultrasound of the abdomen and pelvis revealed no abnormalities.

The diagnosis of intralobar bronchopulmonary sequestration was made based on history and imaging findings. The patient was managed conservatively with antibiotics and tranexamic acid and was subsequently reviewed by cardiothoracic surgeons who advised outpatient review after completion of antibiotics.

The patient had video-assisted thoracoscopic surgery (VATS) resection of the sequestered lung with good postoperative recovery.

Follow-up chest X-ray showed low lung volume compatible with left lower lobe resection without the presence of the previous consolidation (figure 7).

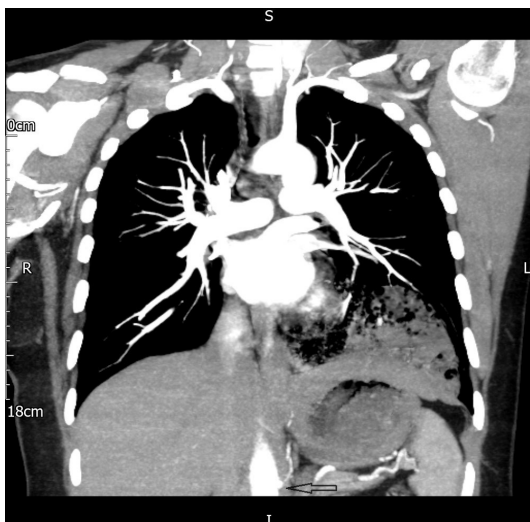


Figure 4 CT of the chest (mediastinal window) with vessel origin from coeliac plexus.

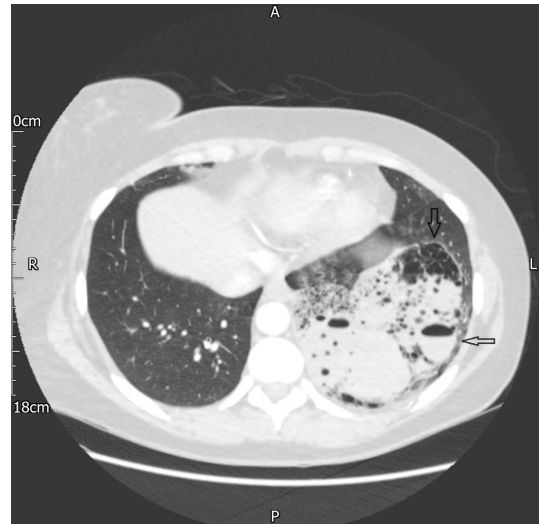


Figure 5 CT of the chest (axial view) showing multiple cystic lesions.

Histopathology of the resected segment confirmed the diagnosis of intralobar pulmonary sequestration.

INVESTIGATIONS

CT of the chest with contrast and chest X-ray (preoperative and postoperative).

Ultrasound of the abdomen and pelvis.

DIFFERENTIAL DIAGNOSIS

Congenital diaphragmatic hernia.

Mediastinal tumours such as teratoma and neuroblastoma.

Bronchogenic cyst.

TREATMENT

The patient had left-sided VATS to remove the sequestered left lower lobe of the lung.

Features consistent with intralobar sequestration were confirmed on postoperative histopathology specimen.

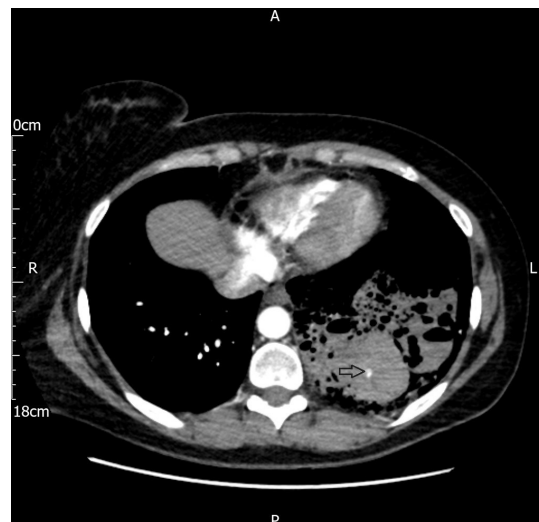


Figure 6 CT of the chest (mediastinal window) demonstrating feeding vessel with multiple cystic lesions.

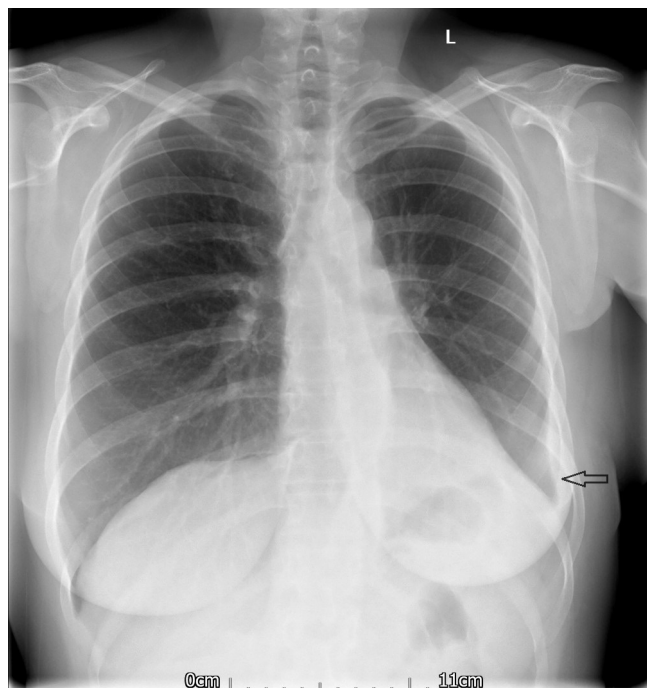


Figure 7 Postoperative chest X-ray showed low lung volume compatible with left lower lobe resection without the presence of the previous consolidation.

OUTCOME AND FOLLOW-UP

No immediate postoperative complications were noted. The patient was discharged with advice to follow-up in 3 months with a repeat chest X-ray.

DISCUSSION

Intralobar pulmonary sequestration is a relatively rare congenital anomaly with few reports of initial diagnosis occurring during adulthood. The most common location of intralobar sequestration is in the posterior basal segments of the lung, with nearly two-thirds appearing in the left lung. Associated congenital anomalies are uncommon in intralobar sequestration.⁵ Some patients are asymptomatic, whereas others may present with persistent cough, expectoration and haemoptysis.

Diagnosis is usually made with imaging such as CT of the chest and in some patients may need angiography.⁶ In intralobar sequestration, the anomalous systemic arterial supply is via the descending thoracic aorta in 72% of cases, from the abdominal aorta and its branches in 21% of cases, from intercostal artery in 3% of cases, and rarely from the subclavian, internal thoracic and pericardiophrenic arteries.⁷ In our case, the blood supply was from the coeliac axis which is rare.

CT of the chest effectively diagnosed the condition and also delineated the anatomy in this case, which was useful in surgical resection.

A chest X-ray can provide a diagnostic clue to lung sequestration. In the past, aortography was frequently used for diagnosis. However, the gold standard for identifying the pulmonary

sequestration is CT/MR angiography as it confirms the anatomy and also identifies the anomalous systemic arterial supply and the venous drainage.

An asymptomatic patient can be observed, while those with recurrent symptoms require surgical resection of sequestered segments.

Other treatment modalities include arterial embolisation, especially in a lesion with a single well-characterised systemic arterial supply with high-output cardiac failure.⁸

Learning points

- ▶ Diagnosing a case of adult congenital malformations can be challenging. Therefore, one should consider a wide differential diagnosis for evaluating a patient with unexplained haemoptysis.
- ▶ The feeding vessel from the coeliac axis can only be demonstrated through contrast-enhanced CT, and thus when in doubt consider angiography by contrast-enhanced imaging or in rare cases by invasive angiography.
- ▶ Timely diagnosis and treatment will not only minimise the patient's distress but also decrease the burden on the healthcare system.

Contributors DMS contributed by referring the patient to cardiothoracic surgeons and also in writing the abstract. AA contributed by taking care of the patient while in ward and discussing with radiologist for the diagnosis. UD contributed in referrals and writing the case sheet. NS, the corresponding author, contributed in most of the steps, including diagnosing, discussing and convincing the radiologist about the diagnosis and taking patient consent form.

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